

*Focused on Improving the Lives of Patients with Rare Genetic Mitochondrial Diseases*



**UMDF  
Clinical Trial Update  
June 30, 2023**

# Forward-Looking Statements

This presentation contains "forward-looking" statements that are based on our management's beliefs and assumptions and on information currently available to management. Forward-looking statements include all statements other than statements of historical fact contained in this presentation, including information concerning our business strategy, objectives and opportunities. Forward-looking statements are subject to known and unknown risks, uncertainties, assumptions and other factors that may cause our actual results, performance or achievements to differ materially and adversely from those anticipated or implied by our forward-looking statements, including, but not limited to, those related to our dependence on third-party clinical research organizations, manufacturers, suppliers and distributors; our ability to obtain necessary additional capital; our ability to obtain necessary regulatory approvals for our products and, if and when approved, market acceptance of our products; the commercialization plans and expectations for commercializing mavodelpar (REN001) in the United States and rest of world, estimates of the number of patients impacted by PMM or LC-FAOD and who are appropriate for treatment with mavodelpar, the potential benefits of mavodelpar, the financial impact or revenues from any commercialization we undertake, the impact of competitive products and therapies; our ability to attract and retain key employees; the costs of our commercialization plans and development programs; the design, implementation and outcomes of our clinical trials; our ability to manage the growth and complexity of our organization; our ability to maintain, protect and enhance our intellectual property; and our ability to continue to stay in compliance with applicable laws and regulations. You should refer to the section entitled "Risk Factors" set forth in our Annual Report on Form 10-K, Quarterly Reports on Form 10-Q and other filings we make with the Securities and Exchange Commission (SEC) from time to time for a discussion of important factors that may cause our actual results to differ materially from those expressed or implied by our forward-looking statements. You should not rely upon forward-looking statements as predictions of future events. Neither we nor any other person assumes responsibility for the accuracy and completeness of the forward-looking statements. We undertake no obligation to update any forward-looking statements after the date of this presentation except as may be required by law.

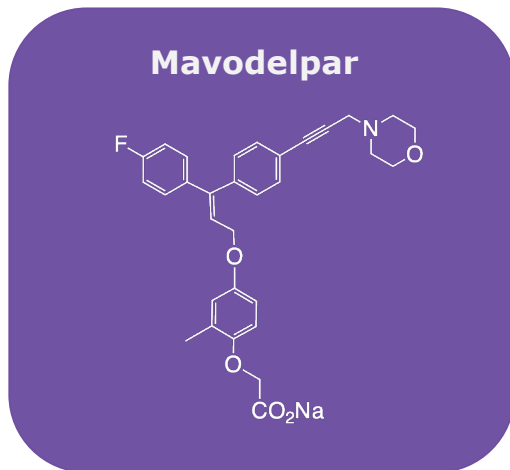
This presentation also contains estimates and other statistical data made by independent parties and by us relating to market size and growth and other data about our industry. These data involve a number of assumptions and limitations, and you are cautioned not to give undue weight to such estimates. Projections, assumptions and estimates of the future performance of the markets in which we operate are necessarily subject to a high degree of uncertainty and risk. The trademarks included herein are the property of the owners thereof and are used for reference purposes only.

Mavodelpar is an investigational drug product candidate that is under clinical investigation, and which has not yet been approved for marketing by the U.S. Food and Drug Administration, European Medicines Agency, or any other global regulatory agency. No representation is made as to the safety or effectiveness of this product candidate for the use for which it is being studied.

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# Mavodelpar is a Selective PPAR Delta (PPAR $\delta$ ) Agonist

Mavodelpar is a once-daily oral drug in development for patients with Primary Mitochondrial Myopathies (PMM)

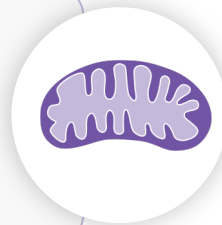


Mavodelpar is an investigational drug that is not authorized for marketing or sale in any country at the time of this presentation

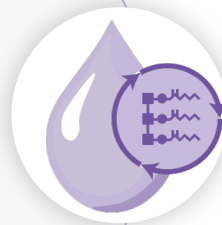
1. Increases transcription of genes central to mitochondrial function



2. Drives production of new mitochondria

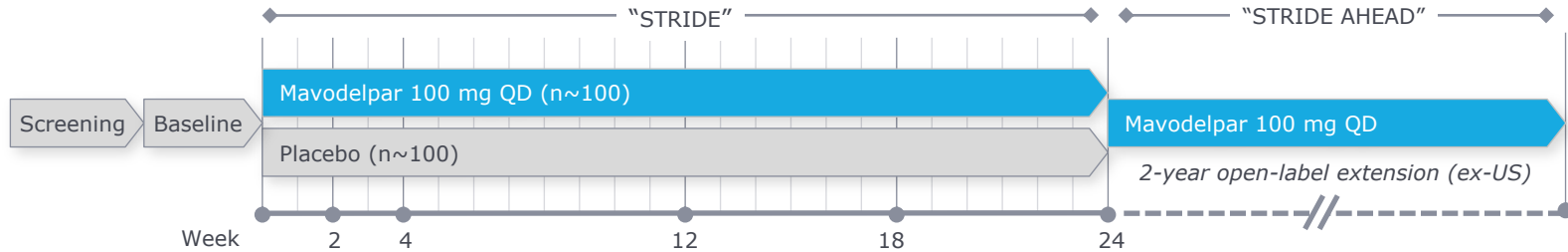


3. Increases oxidation of fatty acids and cellular energy production



# Pivotal PMM Phase 2b (STRIDE) Study: Overview

**Randomized, double-blind, placebo-controlled clinical trial in adults with mtDNA defects and myopathy**



## Primary Endpoint

- Change from baseline to week 24 in distance walked during 12MWT

## Secondary and Exploratory Endpoints

- Changes from baseline in PROMIS® Short Form Fatigue 13a, Modified Fatigue Impact Scale (MFIS), Patient Global Impression of Change (PGIC), Patient Global Impression of Severity (PGIS), 30 Second Sit-To-Stand (30STS) Test, Brief Pain Inventory (BPI), 36-Item Health Survey (SF-36), Work Productivity and Activity Impairment Questionnaire: Specific Health Problem (WPAI:SHP), and Pedometer Step Counts

# Reneo's PMM Program Update

- We have completed enrollment (N=213 patients) in the pivotal STRIDE study of mavodelpar (REN001) in adult patients with PMM
  - *Topline results anticipated in by the end of 2023*
- We have enrolled over 120 patients in STRIDE AHEAD, the mavodelpar open-label extension (OLE) study in adult patients with PMM
  - *Around 75 patients have been treated for more than 6 months in the study*
- The STRIDE AHEAD study was amended to allow the enrollment of adult patients with PMM due to nuclear DNA (nDNA) defects
  - *We expect first nDNA patients to enroll soon*
- If successful, the results of the STRIDE and STRIDE AHEAD studies will form the basis for Marketing Applications to the FDA, EMA and other regulatory agencies

To learn more, please visit:  
*[ClinicalTrials.gov](https://clinicaltrials.gov)*

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We would like to thank the PMM  
community - especially the  
patients who have participated  
in our studies - for their support

Alejandro Dorenbaum, MD

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*Reneo Pharmaceuticals, Inc.*